

## CASE REPORT

## Kaposi's sarcoma infiltrating skeletal muscle

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An HIV-1 antibody positive black African man with plasma cell variant Castleman's disease and cutaneous Kaposi's sarcoma, despite receiving chemotherapy, had progressive disease. In addition, he developed pain and swelling behind the right knee. Histology of an ultrasound guided biopsy showed Kaposi's sarcoma infiltrating the head of gastrocnemius.

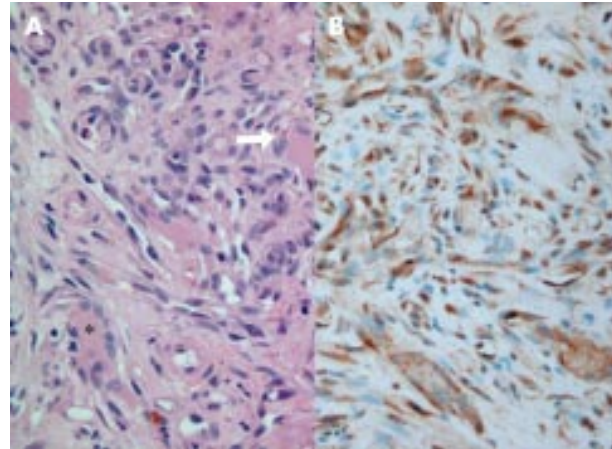
A 32 year old black African man was found to be HIV-1 antibody positive in June 2001, when he presented with widespread lymphadenopathy, splenomegaly, and cutaneous Kaposi's sarcoma. Investigations revealed a pancytopenia; histology of an axillary lymph node showed plasma cell variant multicentric Castleman's disease. Antiretroviral therapy with lopinavir/ritonavir, abacavir, and lamivudine was commenced. Chemotherapy with CHOP (cyclophosphamide, adriamycin, vincristine, and prednisolone) was poorly tolerated and after two cycles was modified to vincristine and prednisolone only, of which he received four cycles.

The patient presented in early January 2002 with increasing pain behind the right knee and slight swelling of the right ankle. Examination in addition revealed new cutaneous Kaposi's sarcoma lesions on the right ankle and shin. Ultrasound did not show deep vein thrombosis; a 1.5 cm lymph node was noted in the right popliteal fossa.

He re-presented 3 weeks later with persistent pain behind the right knee, a non-productive cough, loss of appetite, and fever. Examination showed hepatosplenomegaly. Investigations showed haemoglobin 7.4 g/dl, white blood cells  $3.2$  (neutrophils  $1.4$ )  $\times 10^9/l$ , platelets  $136 \times 10^9/l$ . A bone marrow aspirate and trephine showed only hypercellular, dysplastic changes which were ascribed to HIV. At this time his CD4 count was  $340 \text{ cells} \times 10^6/l$  and viral load was 80 copies/ml. An abdominal computed tomograph (CT) scan showed hepatosplenomegaly, the spleen having enlarged despite chemotherapy. CT pulmonary angiography showed bilateral arterial thrombi, and he was commenced on low molecular weight heparin.

Interferon alfa, 3 MU subcutaneously given three times per week, was started as salvage therapy for the Castleman's disease. Over the next 3 weeks the patient reported increasing pain and swelling behind the right knee. Examination revealed a woody hard mass in the head of the gastrocnemius muscle. This was not fixed to the overlying skin, and there was no Kaposi's sarcoma overlying the area. Ultrasound scanning of the limb showed a soft tissue mass infiltrating the head of gastrocnemius. Histology of an ultrasound guided percutaneous biopsy showed Kaposi's sarcoma infiltrating muscle (fig 1). Treatment with local radiotherapy produced rapid improvement in pain and in the size of the lesion.

This man's case is unique in that Kaposi's sarcoma invading the muscle tissue has not previously been reported. An association between the development of Kaposi's sarcoma and the presence of Castleman's disease has been described in the literature,<sup>1–3</sup> and it has been postulated that Castleman's disease



**Figure 1** (A) Regenerating muscle fibres (asterisk, lower left) are surrounded by tumour. The tumour comprises uniform spindle cells with a fascicular architecture and numerous slit-like vessels containing red blood cells. There are hyaline globules (arrow) and deposition of hemosiderin (not shown). HHV-8 positivity (B) excludes the differential diagnoses of bacillary angiomatosis and spindle cell haemangioendothelioma. The tumour was also positive for CD34 and CD31, and negative for factor VIII (not shown). These are the typical staining characteristics of Kaposi's sarcoma.

is directly implicated in the pathogenesis of Kaposi's sarcoma, although there are no reports of differences in the natural history or aggressiveness of Kaposi's sarcoma when these two conditions coexist. This lesion and new cutaneous lesions developed despite good virological control of HIV. It also developed while on treatment with interferon alfa, which is also effective against Kaposi's sarcoma.<sup>3,4</sup> The lesion did however respond symptomatically to radiotherapy. Our patient had a history of venous thromboembolism, and the diagnosis may have been delayed by the false reassurance that this was not evident on an earlier ultrasound. This case emphasises the importance of biopsy of soft tissue masses in patients with AIDS or with multiple previous pathologies. It is recognised that this condition may easily be confused with bacillary angiomatosis, which has been previously reported as occurring in muscle in HIV positive patients.<sup>5,6</sup> In addition, certain clinicopathological features are shared with spindle cell haemangioendothelioma.<sup>7</sup>

## CONTRIBUTORS

LJH abstracted the clinical and pathological records and wrote the first and final drafts of the manuscript; SD reported on histology of the biopsy, created the images and wrote the figure legend; SB performed ultrasound guided biopsy, reported findings on ultrasound and commented on the drafts of the manuscript; RFM proposed and coordinated the project, cowrote the first and final drafts.

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